SELF ASSESSMENT QUESTIONS

A case of digital vasculitis

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Answers on p 723.

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A 49 year old woman presented in June 1998 with left sided pleuritic chest pain, fever, night sweats, flitting arthralgia, and loss of appetite of three weeks' duration. She also complained of episodic numbness of her fingertips and later her toes and oral ulceration.

She was known to have had asthma for the past 24 years and to have had four episodes of arthralgia involving large joints in the last year.

On general examination, she was pyrexial (37.5°C) with evidence of digital vasculitis affecting the left index and middle finger nails (see figs 1 and 2). She had oral ulceration. There was no rash or any evidence of inflammatory arthritis. Examination of the peripheral nervous system revealed reduced power of her right plantar flexion and inversion of right foot. There was diminished sensation over the sole of the right foot and sensory loss affecting all fingertips.

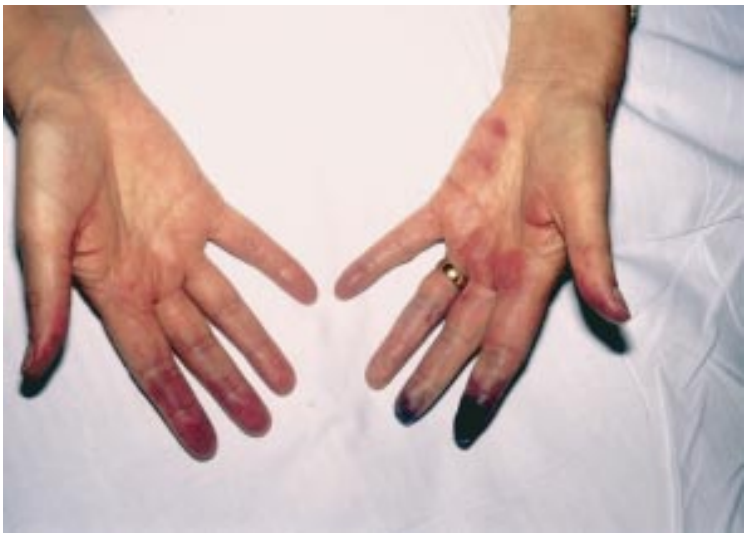

Figure 1 Digital vasculitic lesions.

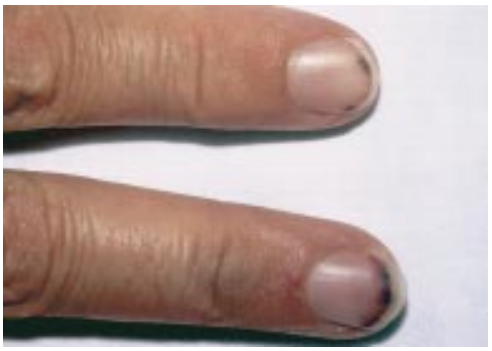

Figure 2 Vasculitic lesions affecting the left index and middle fingers.

Investigations on admission gave the following results: white cell count 21.3 × 10^9/l, eosinophils 5.4 × 10^9/l, neutrophils 13.1 × 10^9/l, haemoglobin 128 × g/l, platelets 200 × 10^9/l, serum C reactive protein 145 mg/l, and erythrocyte sedimention rate 74 mm/hour. Her biochemical profile was normal, except for a moderately raised alkaline phosphatase of 176 IU/l and creatinine kinase of 202 IU/l with normal creatinine kinase myocardial bound isoenzyme.

Chest radiography showed diffuse patchy consolidation of both lungs. Spiral computed tomography of her chest showed a little patchy consolidation of upper lobes suggestive of vasculitis. An echocardiogram was normal.

Urinalysis revaled microhaematuria and proteinuria. A renal biopsy and angiogram were normal. Her latex, antineutrophil cytoplasmic antibodies, antinuclear antibodies, anticardiolipin antibody titres, and hepatitis B surface antigen were all negative. A ceretec scan was normal.

Questions
(1) What is the most probable diagnosis?
(2) What is the treatment?

Abdominal wall thickening in a middle aged man

M Bhushan, R H Macdonald, M H Irving, C E M Griffiths

Answers on p 723.

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A 44 year old man presented to the surgical team with a three week history of a sensation of increasing tightness of the lower abdominal wall. There had been no obvious precipitating factors, clinical prodrome, or associated systemic symptoms. He had been previously well and took no regular medication of note. Initial inspection of the skin was unremarkable but palpation revealed a firm, non-tender induration of the skin (11 × 4.5 cm) around the umbilicus. The rest of the skin was unaffected. General examination was normal. A peripheral blood eosinophilia of 1.5×10^9/l (normal $0.2–0.4 \times 10^9$/l) was noted. All other laboratory investigations including erythrocyte sedimentation rate, blood film, renal and hepatic function, and immunology screen were normal. Computed tomography and magnetic resonance scans of the abdomen were also reported as normal. An open biopsy, down to and including rectus sheath, was performed under general anaesthesia. Histology of the biopsy is shown in fig 1.

Questions
(1) What is the diagnosis?
(2) What is the pathogenesis of this condition?

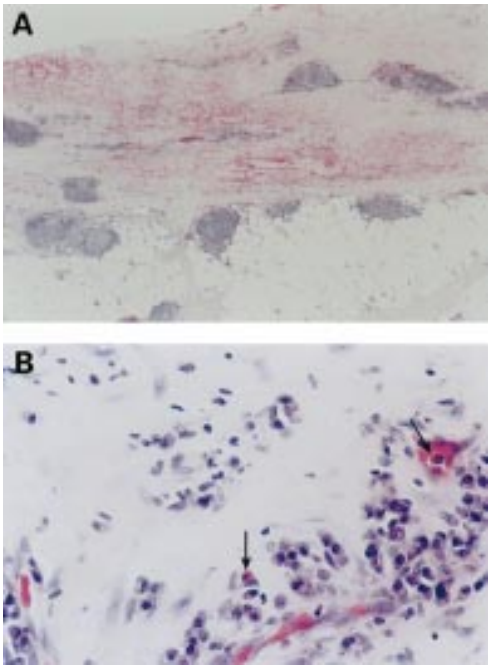

Figure 1 Histology of the skin biopsy (haematoxylin and eosin (A) magnification × 25 and (B) magnification × 400).

A 71 year old man with a facial lesion

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Answers on p 724.

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Submitted 27 July 1999
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A 71 year old man (fig 1) was brought into the accident and emergency department by his two daughters. He had had a lesion on the left side of his face for five years and has refused to see a doctor. The lesion had become progressive over the past year and he was finally persuaded to seek medical attention.

Questions
(1) What is the diagnosis?
(2) Discuss treatment for this condition

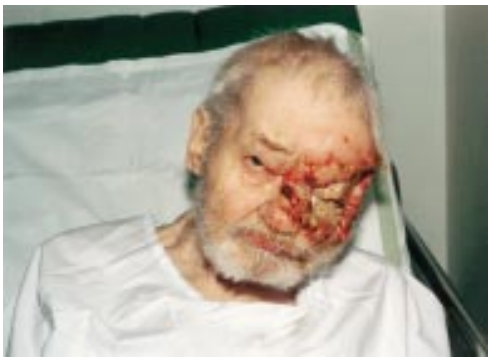

Figure 1 Facial lesion (reproduced with the patient's permission).

The pale and limping child

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Answers on p 725.

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A 13 year old girl presented with a two month history of recurrent episodes of arthritis of the metacarpophalangeal, wrist, knee, ankle, and metatarsophalangeal joints. The arthritis seemed to follow a migratory and symmetric pattern. The arthritis lasted for a week, resolved spontaneously, only to recur a few days later in another joint. Apart from having intermittent low grade fever, she was well. There was no history of sore throat, diarrhoea, or rash. On examination she was pyrexial (temperature 37.6°C). There was pallor, hepatomegaly (liver span measuring 17 cm), and moderate splenomegaly. The left and right first metacarpophalangeal joints were swollen and tender while there were residual swellings of the ankles. Initial investigations revealed a haemoglobin of 89 g/l, leucocyte count of 7.3×10^9/l with normal differential count and platelet count of 82×10^9/l. Erythrocyte sedimentation rate was 125 mm/hour. Levels of complement and immunoglobulins were normal. Rheumatoid factor, HLA B27, antinuclear, anti-double stranded DNA, and antistreptolysin O antibodies were negative. Culture results were unremarkable. The patient received non-steroidal anti-inflammatory drugs and two units of red cells. Two weeks later she returned with worsening of the arthritis and generalised lymphadenopathy. The platelet count dropped to 16×10^9/l. Peripheral blood and bone marrow smears are shown in fig 1A and B respectively.

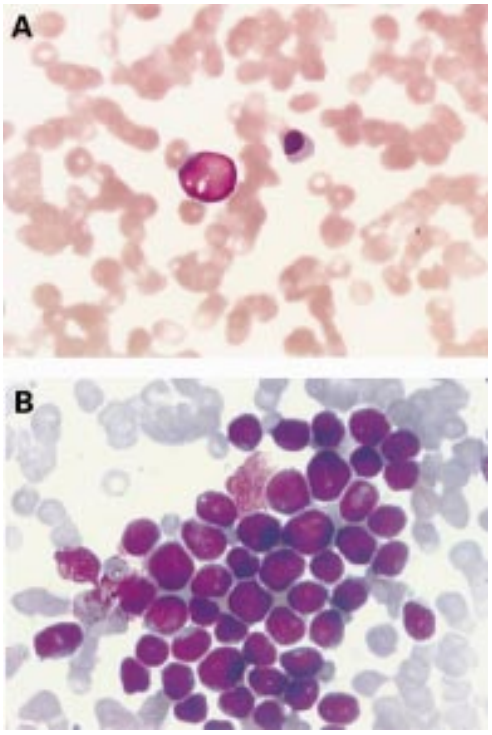

Figure 1 Smears of the peripheral blood (A) and bone marrow aspirate (B).

Questions
(1) What are the differential diagnoses of the arthritis?
(2) What do the peripheral blood and bone marrow smears show?

Rigidity, hyperthermia, and altered mental status

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Answers on p 726.

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A 55 year old white woman presented with a two day history of severe rigidity, hyperthermia, altered mental status, and autonomic dysfunction. She had been treated for paranoid-type schizophrenia with zuclopentixol for two years. Thirteen weeks before, zuclopentixol had been discontinued and she started treatment with olanzapine (10 mg/day). Two weeks before, olanzapine was stopped because of worsening of her schizophrenic symptoms and she started taking zuclopentixol again (30 mg/day). She did not use any other medications or drugs. The clinical picture happened in winter. On admission, the oral temperature was 40.3°C, blood pressure 110/70 mm Hg, pulse rate 125 beats/min, and respiratory rate was 30/min. There was prominent diaphoresis. The neurological examination was difficult to complete because of the patient's uncooperativeness. She did not follow commands or answer questions. His face was symmetric, and she had severe rigidity in neck, arms, and legs. Deep tendon reflexes were symmetric and the results of the Babinski test were equivocal. Extraocular movements and fundi were normal.

Questions
(1) What is your differential diagnosis for this case and what investigations would you perform?
(2) What is the treatment for this case?

A rare cause of recurrent meleana in an elderly women

B Spencer, A Akpan, D King

Answers on p 727.

A 77 year old woman with a medical history of peptic ulcer disease and irritable bowel syndrome presented with an *Escherichia coli* septicaemia secondary to a urinary tract infection, which was treated successfully with intravenous cephalosporins. During her recovery she had an episode of melaena during which her haemoglobin dropped to 67g/l from within the normal range. She was transfused, and was haemodynamically stable. Gastroscopy revealed atrophic gastritis, colonoscopy was normal to the splenic flexure, and a barium enema revealed diverticular disease. She had no further bleeding and was discharged. Four months later she was admitted with a further bleed, she was haemodynamically stable but her haemoglobin had dropped 30 g/l to 88 g/l. At this stage a diagnosis of angiodysplasia was considered but as she was no longer actively bleeding mesenteric angiography was not undertaken. She was transfused and discharged. Three months later she had a third episode of melaena, at this point further gastrointestinal imaging was performed.

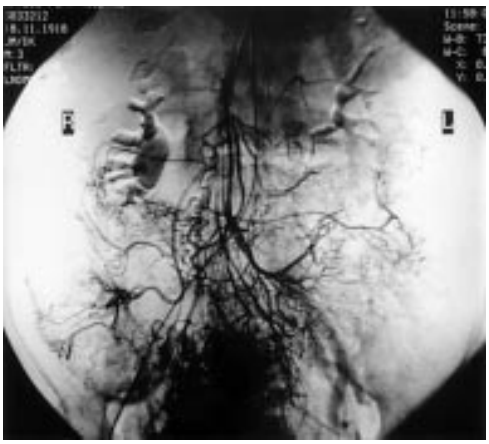

Figure 1 Mesenteric arteriogram.

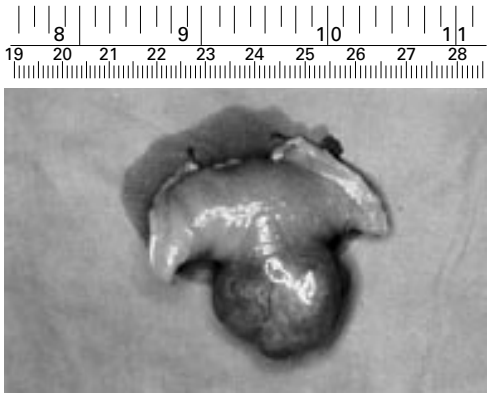

Figure 2 Laparotomy specimen.

At laparotomy there was a small 4 × 2 cm polypoid lesion on the serosal surface of the mid-small bowel. Subsequent histological examination revealed a tumour with a low mitotic rate, composed of both smooth muscle spindles and pancreatic glandular elements; this was consistent with a benign small bowel myoepithelial hamartoma. The patient has remained well and has had no further bleeds.

Questions

(1) What is meant by the term "obscure gastrointestinal haemorrhage" and how would you investigate it?
(2) What is shown in fig 1?
(3) Figure 2 shows a laparotomy specimen consisting of a 4 × 2 cm polypoid lesion on the serosal surface of the mid-small bowel. Histologically this is a myoepithelial hamartoma. What are hamartomas?

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Abdominal lump in an infertile man

Answers on p 728.

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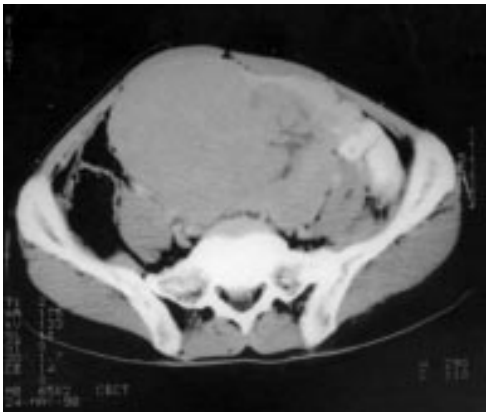

Figure 1 Contrast enhanced computed tomogram at the level of lower abdomen showing a bulky enhanced mass.

Figure 2 Cut section of the tumour is solid, homogenous and grey-white in colour.

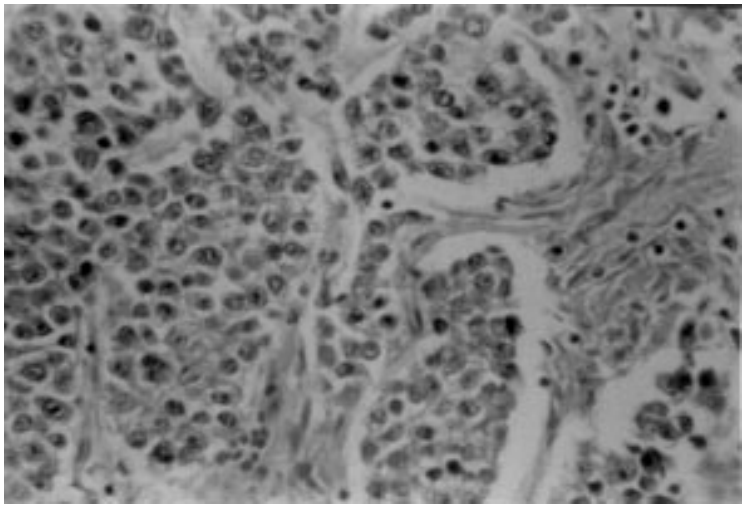

Figure 3 Lobules of monotonous tumour cells with clear cytoplasm and centrally located vesicular nucleus (haematoxylin and eosin, magnification × 400).

A 44 year sexually active man presented with pain in his abdomen, dysuria, haematuria, constipation, and a lump in the suprapubic area of over four months' duration. Further exploration into his medical history revealed that he had been married for 20 years but had no children and had been diagnosed as having primary infertility. Examination showed a man of average build, with a mass in the lower abdomen. The intra-abdominal mass measured 12 × 12 cm and spanned from the umbilicus to pubis vertically and from left flank to across the midline horizontally. The consistency of the lump was firm to hard, non-tender, and fixed to underlying structures. Distension of the large bowel was evident above the mass. There was no testicle in the left scrotum and a small right testicle in the groin.

With a presumptive diagnosis of pelvic malignancy, complicated with intestinal obstruction, radiography of the abdomen was undertaken and this showed evidence of subacute intestinal obstruction. Gastrograffin enema *x* rays revealed a soft tissue mass adherent to the rectosigmoid junction suggestive of carcinoma of the urinary bladder. Subsequent contrast enhanced computed tomography of the abdomen and pelvis revealed an isodense soft tissue mass with areas of hypodensity arising from pelvis measuring 12 × 12 × 12 cm with minimal peripheral enhancement (fig 1). There was no definite cleavage between tumour and the bladder wall. Serum α-fetoprotein and β-human chorionic gonadotrophin concentrations were within normal limits.

An explorative laparotomy was performed which revealed a large cystic tumour arising from retropubic space of the pelvis pushing the sigmoid colon up and right. The above tumour was measuring 10 × 16 cm in size with distended venous markings overlying. The tumour weighed 1.68 kg. Cut section of the mass showed a grey-white lobular mass with little areas of necrosis (fig 2). There were multiple lymph nodes in the mesentery and paraortic area. Microscopic examination showed uniform looking cells arranged in nests with intervening delicate fibrovascular stroma. The tumour cells were large and have clear cytoplasms. Periodic acid schiff stain for glycogen was strongly positive. The nuclei were centrally located, vesicular, and hyperchromatic, showing numerous mitotic figures (21 per 10 high power field) (fig 3). The sampled tissue from sigmoid colon, bladder, mesenteric and paraotic lymph nodes showed tumour infiltration.

Questions

(1) What is the probable diagnosis of this condition?
(2) Which is the best treatment in this situation?
(3) What is the chance of second cancer in the opposite testis?

A vanishing pituitary mass

Answers on p 729.

N Norman Chan

A 26 year old music composer presented with sudden onset of frontal headache followed by an episode of witnessed tonic-clonic convulsion which lasted 10 minutes. He had bitten his tongue and was confused for 20 minutes. There was no visual disturbances. He was previously in good health without a history of epilepsy or other illnesses. There was no family history of epilepsy. His alcohol intake had been 5–10 units per week for the past eight years.

The patient had a normal BM of 6.8 when checked by the ambulance crew. On arrival in the accident and emergency department, physical examination was unremarkable. The patient became more alert and his Glasgow coma scale score was 15/15. His vital signs were normal with a blood pressure of 150/84 mm Hg. There was no focal neurology or signs of meningism. Fundoscopy was normal and visual field was full on direct confrontation. Blood tests including blood glucose (5.0 mmol/l), electrolytes, liver function, and full blood count were all normal. A magnetic resonance imaging (MRI) scan of the skull was performed (fig 1, left). He did not receive any treatment and a repeat MRI scan was performed seven months later (fig 1, right).

Questions

(1) Describe the initial abnormality shown by the MRI scan (fig 1, left).
(2) What does the follow up MRI scan show (fig 1, right)?
(3) What is the most likely diagnosis?

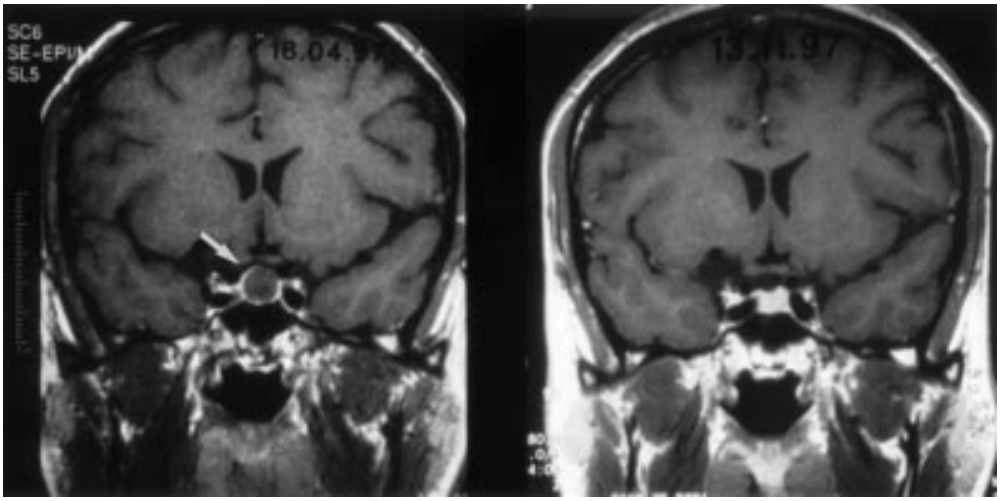

Figure 1 MRI scan on admission (left) and after seven months (right).

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Shortness of breath and diffuse chest pain

Answers on p 731.

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A 60 year old man presented to the emergency medicine department of Sher-i-Kashmir Institute of Medical Sciences, Srinagar with a one day history of shortness of breath and diffuse chest pain aggravated by breathing. He had no history of trauma, fever, altered sensorium, syncope, cough, haemoptysis, weakness, or oliguria. He had a three month history of generalised aches and pains and easy fatigability for which he had received non-steroidal anti-inflammatory drugs and was not evaluated.

Clinical examination revealed moderate pallor, tachycardia, tachypnoea, a depressed anterior chest wall with sharp indentations around the mid-clavicular line on both sides, paradoxical motion of the anterior chest wall, and diffuse bone tenderness. There was no cyanosis, oedema, lymphadenopathy, or organomegaly, and cardiovascular and neurological variables were normal.

Preliminary investigations revealed a haemoglobin concentration of 80 g/l, a normocytic normochromic peripheral smear, total leucocytic count 5.5×10^9/l, and platelet count 200×10^9/l. Erythrocyte sedimentation rate was 65 mm/hour (Wintrobe's), serum urea nitrogen 26.5 mmol/l, creatinine 194.5 μmol/l, calcium 2.9 mmol/l with a normal blood glucose, electrolytes (sodium, potassium), liver profile, alkaline phosphatase, and routine urine examination. Chest radiography revealed double fractures in the 4th, 5th, 6th, and 7th ribs and osteoporosis. Arterial blood gas analysis showed a pH of 7.45, carbon dioxide tension 4.67 kPa, oxygen tension 9.33 kPa, and bicarbonate 19 mmol/l with a saturation of 90%. Further evaluation of the patient revealed presence of Bence Jones protein in the urine (Kappa), presence of M band (quality not determined) on serum and urine electrophoresis, and serum immunoglobulin concentrations of IgG 36 g/l (normal 8–15 g/l), IgA 1.3 g/l (0.9–3.2 g/l), and IgM 0.8 g/l (0.45–1.5 g/l). A skeletal survey revealed multiple lytic lesions in the skull, diffuse osteoporosis, and compression fracture at T4, T5, L4, and L5 vertebrae. Bone marrow examination revealed 35% plasmacytosis.

Questions
(1) What is the diagnosis?
(2) What is the primary disease?
(3) What are the causes of flail chest?
(4) What are the treatment options?

Answers on p 731.

An elderly man with muscle cramps

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A 60 year old man presented to the emergency department with slurring of speech and generalised weakness associated with cramps affecting his hands. He had no medical illnesses apart from a left sided cataract removed a year ago. A 12 lead electrocardiogram was performed.

Questions
(1) What is the abnormality on the 12 lead electrocardiogram (fig 1)?
(2) What are the possible causes of the abnormality of the electrocardiogram?
(3) What is the likely diagnosis?

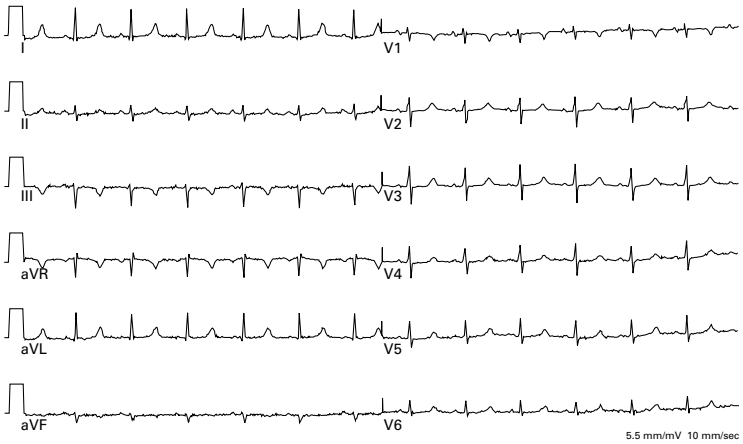

Figure 1 Electrocardiogram.

Left kidney mass in a 45 year old woman

Answers on p 733.

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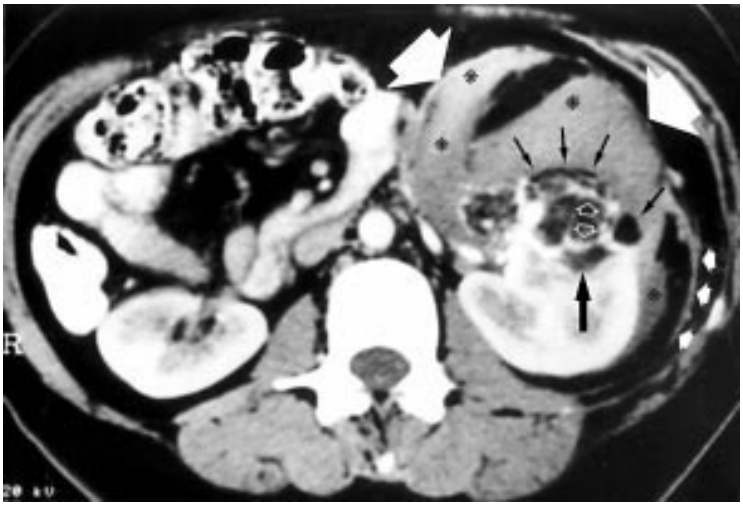

Figure 1 Abdominal contrast enhanced computed tomogram.

A 45 year old woman presented with left sided flank pain, anaemia, and severe hypotension. On clinical examination, a subcutaneous mass was palpated in the left flank and hypocondrium.

Renal urography showed the presence of a large mass with dislocation and compression of left kidney pelvis. Abdominal ultrasound revealed a highly reflective lesion in the left kidney. Abdominal contrast enhanced computed tomography was performed (fig 1).

Questions

(1) What is the most likely diagnosis?
(2) What is the treatment?

Accepted 8 October 1999

SELF ASSESSMENT ANSWERS

A case of digital vasculitis

Q1: What is the most probable diagnosis?
The diagnosis is Churg-Strauss syndrome on the basis of digital vasculitis, Raynaud's phenomenon, mild peripheral neuropathy, mononeuritis multiplex, marked eosinophilia, and a past history of asthma.

Q2: What is the treatment?
The treatment is steroids. Patients with poor prognostic features, such as myocardial and gastrointestinal involvement, can be treated with combined steroids and cyclophosphamide.

Discussion
Churg-Strauss syndrome (CSS) is a granulomatous vasculitis affecting multiple organ systems. It may resemble polyarteritis nodosa except for the high degree of lung involvement, vasculitis of mainly small sized vessels, extravascular granuloma formation, eosinophilic tissue infiltration, peripheral eosinophilia, and a strong association with severe asthma.[1]

The American College of Rheumatology has proposed six criteria for the diagnosis of CSS. Four of the six criteria are necessary for a diagnosis with 85% sensitivity and 99.7% specificity. These criteria include a history of asthma, the presence of eosinophilia >1500/mm^3, paranasal sinusitis, pulmonary infiltrates seen on radiography, histological proof of vasculitis, and mononeuritis multiplex.[2]

Asthma is the most frequently observed presentation but may precede the development of systemic vasculitis by up to 30 years.[3] Mononeuritis multiplex is the second commonest manifestation. Other common features are fever, malaise, weight loss, fleeting pulmonary infiltrates, arthralgia, and gastrointestinal involvement. Cardiac involvement is seen in one third of patients, with myocardial and endocardial involvement being common. Skin lesions are seen in 70% of cases, usually in the form of purpura, cutaneous and subcutaenous nodules. Glomerular involvement is rare. Gastrointestinal tract involvement is due to mesenteric vasculitis and usually presents with abdominal pain.[3 4]

Although CSS may be readily diagnosed on clinical grounds, histological confirmation should always be sought. The classical picture consists of necrotising vasculitis, eosinophilic tissue infiltration, and extravascular granulomas, but it is only found in a minority of patients and is not pathognomic of CSS.[5]

Antineutrophil cytoplasmic antibodies, especially antimyeloperoxidase, are positive in 60%–75% of patients with CSS.[5]

The prognosis is poor with 25% five year survival if untreated. Treatment with steroids dramatically increases the chance of survival. Patients with poor prognostic features, such as myocardial and gastrointestinal involvement, should be treated with combined steroids and cyclophosphamide.

Our patient had four out of the six American College of Rheumatology criteria, namely asthma, eosinophilia, pulmonary infiltrates, and mononeuritis multiplex. Her cardiac involvement warranted the use of cyclophosphamide in addition to methylprednisolone.

Final diagnosis
Churg-Strauss syndrome.

1 Churg J, Strauss L. Allergic granulomatosis, allergic angiitis and polyarteritis nodosa. *Am J Pathol* 1951;**27**:277–301.
2 Masi AT, Hunder GG, Lie JT, *et al.* The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum* 1990;**33**:1094–100.
3 Chumbley LC, Cohen P, Gayraud M, *et al.* Churg-Strauss syndrome. Clinical study and long term follow-up of 96 patients. *Medicine* 1999;**78**:26–37.
4 Lanham JG, Elkon KB, Pusey CD, *et al.* Systemic vasculitis with asthma and eosinophilia: a clinical approach to the Churg-Strauss syndrome. *Medicine* 1984;**63**:65–81.
5 Reid AJC, Harrison BDW, Watts RA, *et al.* Churg-Strauss syndrome in a district hospital. *Q J Med* 1998;**91**:219–29.

Abdominal wall thickening in a middle aged man

Q1: What is the diagnosis?
The biopsy (see p 716) shows marked thickening of the fascia throughout the subcutaneous compartment associated with a chronic inflammatory infiltrate consisting of lymphocytes and plasma cells. Lymphoid follicle formation is also seen. Scattered eosinophils (arrowed) are noted on higher magnification. There is no vasculitis, panniculitis, or fat necrosis. In view of the tissue eosinophilia and initially raised eosinophil count, the diagnosis is eosinophilic fasciitis. A year after his operation the patient is being managed conservatively. There has been spontaneous resolution of his signs (the affected area now measures 6.5 × 2 cm). His eosinophil count returned to normal within three months of his operation and has remained so since this time.

Q2: What is the pathogenesis of this condition?
Eosinophilic fasciitis is considered to be an immunologically mediated disease. A key point in the history is that approximately half of the patients will describe strenuous physical exertion before development of clinical signs and symptoms. It is hypothesised that such exertion might damage muscle, rendering it antigenic and causing an immune response. Such a response is reflected by the dense chronic inflammatory infiltrate, which may include eosinophils in the early stages of the condition, seen within the fascial compartment. These tissue eosinophils are considered to play a central part in the formation of the fascial fibrosis which is the hallmark of this condition.[1]